Rare disease

Bilateral cystic nephroma with pleuropulmonary blastoma

Anand Kumar Bhardwaj,¹ Prem Dass Sharma,¹ Amit Mittal,² Akshay Sharma¹¹Paediatrics Department, MM Institute of Medical Sciences and Research, Ambala, Haryana, India;²Radiology Department, MM Institute of Medical Sciences and Research, Ambala, Haryana, India**Correspondence to** Dr Anand Kumar Bhardwaj, col.akbhardwaj@gmail.com**Summary**

Cystic nephroma is a rare benign renal neoplasm that is purely cystic and is lined by an epithelium. Bilateral cystic nephromas are even rarer with only a handful cases reported in the literature. A case of a 2-year-old male child who presented with bilateral renal cystic masses later diagnosed as cystic nephromas is presented here. Ultrasound, CT scan and histopathological investigations aided in arriving at the correct diagnosis. The most concerning feature was the presence of a fluid filled cystic mass in the lungs, most probably a pleuropulmonary blastoma which is a rare malignant neoplasm known to be associated with bilateral cystic nephromas. The most common presenting symptoms of cystic nephroma are painless abdominal mass, abdominal or flank pain and haematuria. These tumours usually follow a benign course and nephrectomy alone is curative. A close surveillance of such patients is recommended because of elevated risk of other tumours.

BACKGROUND

Cystic nephroma (CN), also called multilocular cystic nephroma, belongs to a heterogeneous group of paediatric cystic renal tumours.¹ It is a rare non-hereditary benign neoplasm of uncertain aetiology that is purely cystic and is lined by an epithelium and fibrous septa that contains mature tubules.² Bilateral CNs are even rarer and till date, only a few paediatric cases of bilateral CNs have been reported.^{1 3–5}

There are two theories on the origin of CN. A developmental origin explains CN as a form of renal dysplasia related to polycystic kidney disease or a result of maldevelopment of the ureteric bud.⁶ The second theory of neoplastic origin explains CN as a beginning of a continuous spectrum of cystic renal tumours which also includes cystic partially differentiated nephroblastoma through to cystic variants of Wilms tumour at the malignant end.⁶

CASE PRESENTATION

A 2-year-old male child was admitted to our tertiary care teaching hospital with complaints of gradually increasing distension of the abdomen for the past one and a half years and pain in the abdomen since last 1 week. There were no constitutional symptoms such as fever, weight loss, respiratory complaints, nausea, vomiting, diarrhoea and dysuria. Bowel and bladder habits were as usual.

This was a full term normal hospital delivery and there were no birth injuries or asphyxia. Prior medical and developmental history was unremarkable. Developmental milestones were normal for age. There was no family history of tuberculosis or any other congenital or chronic illness.

Clinical examination revealed a pale looking child with hypopigmented, sparse hair. There was no oedema, icterus, cyanosis or lymphadenopathy. He was afebrile with vital parameters within normal limits and expected anthropometric measurements. Abdominal examination revealed a large bosselated mass extending from the right

hypochondrium to the right iliac fossa. The mass was firm and non-tender. The liver and spleen could not be palpated separately. There was no free fluid per abdomen. Hernial sites and genitalia were normal.

A chest examination revealed decreased breath sounds in the basal lung fields bilaterally with occasional crackles. Heart sounds were normal and there were no murmurs.

On the basis of the examination findings, a provisional diagnosis of Wilms tumour was made.

INVESTIGATIONS

Ultrasonography revealed a large multiloculated septated ill defined cystic mass on the right side of the retroperitoneum extending beyond the midline. It was inseparable from the renal parenchyma. A similar mass was visualised on the left side, which was smaller in comparison to the one on the right side. A computerised axial tomography scan was done which confirmed bilateral multiloculated multicystic kidney masses measuring 12 cm x 10 cm x 15 cm on the right side (figure 1) and 4 cm x 4 cm x 4.5 cm on the left side (figure 2). No lymphadenopathy was present. It was clearly demonstrated that the renal tissue was well separated from the masses. Histologically, the masses were confirmed to be multilocular bilateral CNs.

An intriguing associated finding in the CT scans and chest radiograph was the presence of a well defined cavity with free fluid in the lower lobe of the right lung (figure 3). There were no associated signs or symptoms with this fluid filled cavity. The patient's parents were not keen on further treatment and left the hospital against medical advice and have since been lost to follow-up. Although a histological diagnosis of the lung cavity could not be made, it is almost certain that the lung mass is a pleuropulmonary blastoma (PPB).^{4 7} Conversely, the most frequent non-pulmonary abnormality associated with a pleuropulmonary blastoma is a CN. There is a greater than 9% association between PPBs and CNs in the PPB registry.

Figure 1 CT scan showing the dimensions of the right kidney.

Figure 2 CT scan showing the dimensions of the left kidney.

Figure 3 CT scan showing the lung cavity.

DIFFERENTIAL DIAGNOSIS

- CN
- Wilms tumour

TREATMENT

CN is a benign malignant condition and hence nephron sparing surgery is possible. In case of small tumours, nephron sparing surgeries are preferred so that as much as possible, renal function may be preserved. In case of bilateral tumours such as in this case, a bilateral surgery with a complete nephrectomy on the side of greater involvement and nephron sparing heminephrectomy on the side of lesser involvement is preferred.

DISCUSSION

CN was first described in 1892 by Edmunds who called the lesion cystadenoma of the kidney.⁸

It is a rare renal neoplasm. Initially it was thought to only occur unilaterally. Powell *et al* established eight diagnostic criteria for these lesions: unilateral involvement; a solitary lesion which is multilocular; there should be no communication with the renal pelvis or of the cysts with each other; the loculi should be lined by epithelium; intralocular septa should be devoid of renal parenchyma; and lastly if residual renal tissue is present, it should be normal.⁹ Boggs and Kimmelstiel, modified these criteria to include the presence of immature renal tissue in the intervening septa.¹⁰ These criteria were further refined by Joshi and Beckwith

and recently some authors have even proposed the elimination of unilaterality as a diagnostic criteria.^{5 11}

Paediatric CN is most frequent in young boys under the age of 4 years and girls older than 4 years.^{12 13} The most common presenting symptoms are painless abdominal mass, abdominal or flank pain and haematuria.^{2 12} The masses are often large, averaging 8–10 cm in size and single.¹⁴ The mass may grow slowly or may demonstrate increase in size rapidly. These tumours usually follow a benign course and nephrectomy alone is curative. Rare local recurrences may be effectively managed by local radiation or chemotherapy.

Ultrasonography is often the first modality used to evaluate abdominal masses. However, large masses may also be evident on radiographs. These are generally well-circumscribed and discrete lesions consisting entirely of cysts and septae, without solid tumour components. A peripheral crescentic rim of renal collecting system helps to confirm the renal origin of the tumour. On contrast CT scan, CN typically appears as a well-circumscribed, encapsulated multicystic mass with no excretion of contrast agent into the loculi. The contents of the cyst either have an attenuation which is either similar or slightly higher than that of water.¹³ Closely packed and small cystic spaces can sometimes mimic a solid mass.¹²

CN is not seen either in the antenatal or neonatal periods, which helps to differentiate it from other tumours such as cystic mesoblastic nephroma.¹⁵

To conclude, although CNs in children are commonly unilateral, they may also be rarely present bilaterally and need to be differentiated from other cystic renal tumours of childhood. The presence of solid components points towards an alternate diagnosis. Given the bilateral nature of some of the cases, nephron sparing surgery is recommended wherever possible, to preserve native renal function. Because the clinical or imaging features of multilocular cystic renal tumours cannot predict its histological characteristics, surgical nephrectomy or nephron sparing surgery is required for both diagnosis and treatment.¹⁶ A close surveillance of such patients is recommended because of elevated risk of other malignancies, such as gonadal tumours, medulloblastoma, leukaemia, lymphoma, thyroid follicular and papillary carcinoma.⁷

Learning points

- ▶ Although classically described as a unilateral entity, CN has been reported to be bilateral as well.
- ▶ CN is a benign neoplasm of unknown aetiology and presents with abdominal enlargement, haematuria and pain in young children.
- ▶ Nephron sparing surgery should be performed for CN as far as possible.
- ▶ A lung lesion notoriously involved with bilateral CN is pleuropulmonary blastoma. It is a malignant lesion and should be actively sought and treated on priority.

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Competing interests None.

Patient consent Obtained.

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